



TITLE:

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CITATION:

MAEKAWA, TAKEO ...[et al]. <Case Report>A Case of Fibrous Histiocytoma of the Liver. 日本外科宝函 1999, 68(1): 14-23

ISSUE DATE:

1999-03-01

URL:

<http://hdl.handle.net/2433/202540>

RIGHT:

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A Case of Fibrous Histiocytoma of the Liver

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Received for Publication, Dec. 4., 1998

Abstract

Malignant fibrous histiocytoma (MFH) is a rare disease. We describe a 68-year-old man admitted to the hospital because of malaise. On admission, hematologic and serum chemical examinations showed no abnormalities. A tumor measuring $6.0 \times 6.0 \times 5.5$ cm was found in segment S6 of the right lobe of the liver. A computed tomographic scan of the abdomen revealed a mass surrounded by a capsule-like region with a nonuniform shadow at its margin. The mass contained a nonuniform low density area. A magnetic resonance imaging scan showed low intensity on T1-weighted images and high intensity on T2-weighted images. An angiogram of the abdomen revealed a tumor with a darkly stained margin during the venous phase. Partial resection of the liver, including S6 and part of S7, was performed. On histopathological examination, this case was characterized by a storiform pattern. The inside of the tumor showed a storiform-pleomorphic pattern with inflammatory cell infiltration and partial mucinous degeneration. On immunohistochemical studies, the tumor cells stained positively for CD6. The diagnosis was MFH.

Introduction

Malignant fibrous histiocytoma (MFH) is a histiocytic tumor derived from fibrous tissue. This tumor rarely arises in the liver. Here we describe our experience with a man undergoing resection for MFH arising in the liver and review the related literature.

Case Report

The patient was a 68-year-old man with malaise. His family history and past disease history were noncontributory. The patient had malaise from about January 1996. In August of the same year, liver dysfunction was diagnosed on a routine health check-up, and he consulted a local physi-

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索引用語: 悪性繊維性組織球腫, 肝臓, 肝切除

Key words: Malignant fibrous histiocytoma, Liver, Hepatectomy

cian. A computed tomographic (CT) scan of the abdomen showed an abnormality in the liver. The patient was introduced and admitted to our department for further evaluation.

On admission, the patient was found to be of moderate build and well nourished. There was no evidence of anemia or jaundice. The blood pressure was 106/50 mmHg, and the pulse rate was 60 beats/min. Physical examination revealed no abnormalities of the chest or abdomen. General hematological examinations showed no evidence of anemia, and serum chemistry revealed no particular abnormalities. Tumor markers (AFP, CEA, CA19-9) were normal. HBs-Ag was negative, but HCV-Ab was positive. ICGR15 was elevated (16.2%) (Table 1).

Echographic examination of the abdomen revealed a mosaic mass measuring 5.3×4.6 cm in S6 of the liver. Immediately superior, there was a low echoic mass measuring 1.5×1.8 cm (Fig. 1). A CT scan of the abdomen disclosed a poorly demarcated low-density area about 50 mm in diameter with a nonuniform interior in the posterior segment of the right lobe of the liver. A capsule-like structure with a heterogenous shadow was seen at its margin (Fig. 2).

Magnetic resonance imaging (MRI) disclosed a mass measuring about 50 mm in diameter in S6. The mass showed low intensity on T1-weighted images and high intensity on T2-weighted images; its center showed higher intensity (Fig. 3). Abdominal angiography showed no distinct vascular abnormalities during the arterial phase, but a darkly stained tumor was seen in S6 during the venous phase (Fig. 4). Although a definitive diagnosis could not be established, hepatocellular carcinoma was suspected, and operation was performed on February 7, 1997.

Table 1 Laboratory data on admission

WBC	8700/mm ³	Glu	111mg/dl
RBC	432×10^4 /mm ³	BUN	15.3mg/dl
Hb	14.3g/dl	Cr	0.7mg/dl
Ht	43.6%	Na	140meq/l
PLT	14.8×10^4 /mm ³	K	5.3meq/l
		Cl	104meq/l
TP	7.7g/dl	AFP	5ng/ml
Alb	3.5g/dl	CEA	2.7ng/ml
T-Bil	0.6mg/dl	CA19-9	16U/ml
D-Bil	0.3mg/dl	ICG R ¹⁵	16.2%
ALP	211IU/l	HBsAg	(-)
GOT	30IU/l	HCVAb	(+)
GPT	29IU/l	PT	29.7sec
LDH	392IU/l	TT	11.1sec
γ -GTP	55IU/l	HPT	65%
ChE	212IU/l		

Thoracolaparotomy was performed via an incision at the right seventh rib. There was no evidence of ascites, peritoneal seeding, or inflammation in the peritoneal cavity. A mass was seen at the posterior segment of the right lobe of the liver. Macroscopically, the liver surface showed findings of moderate cirrhosis. Intraoperative echography showed a tumor located primarily in S6; another tumor with a heterogenous, low echo and measuring 10×15 mm was seen in S7. Liver resection, including S6 and part of S7, was performed.

On histopathological examination, the resected portion of the liver (S6, S7) measured $8.0 \times 7.0 \times 7.0$ cm and contained a tumor measuring $6.0 \times 6.0 \times 5.5$ cm. The surface of a cut section of the tumor was grayish white and parenchymal. A fibrous pseudomembrane was present at the b



Fig. 1 Echographic findings of the liver. A mosaic mass measuring 5.3×4.6 cm was seen in S6 of the liver. Directly superior, a low echoic mass measuring 1.5×1.0 cm was seen.

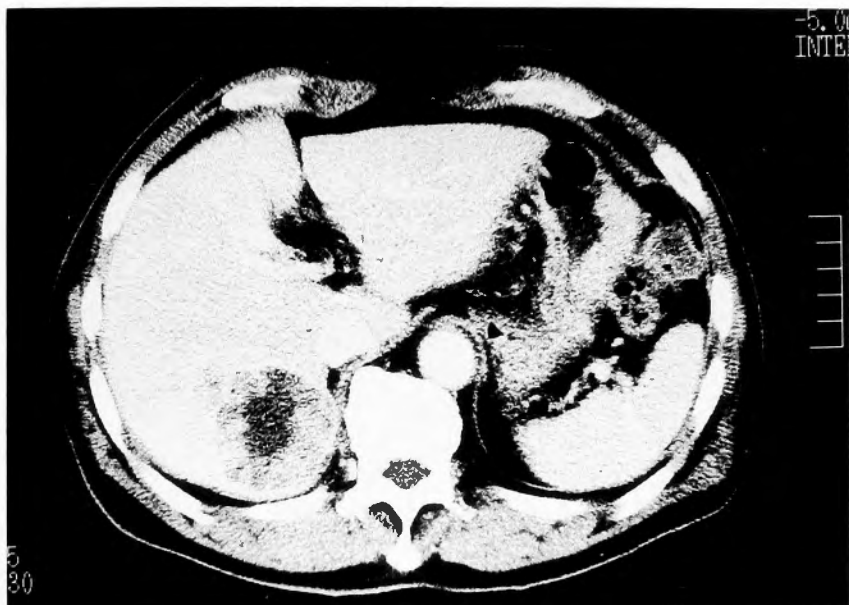


Fig. 2 Abdominal computed tomographic findings. The posterior segment of the right lobe of the liver contained a unclearly demarcated, heterogenous, low density area about 50 mm in diameter. A capsule-like structure with a heterogenous shadow was seen at its margin.

order with the surrounding liver tissue. The tumor contained sporadic hemorrhagic and necrotic foci (Fig. 5). Histologically, the tumor consisted of elongated, spindle-shaped tumor cells proliferat-

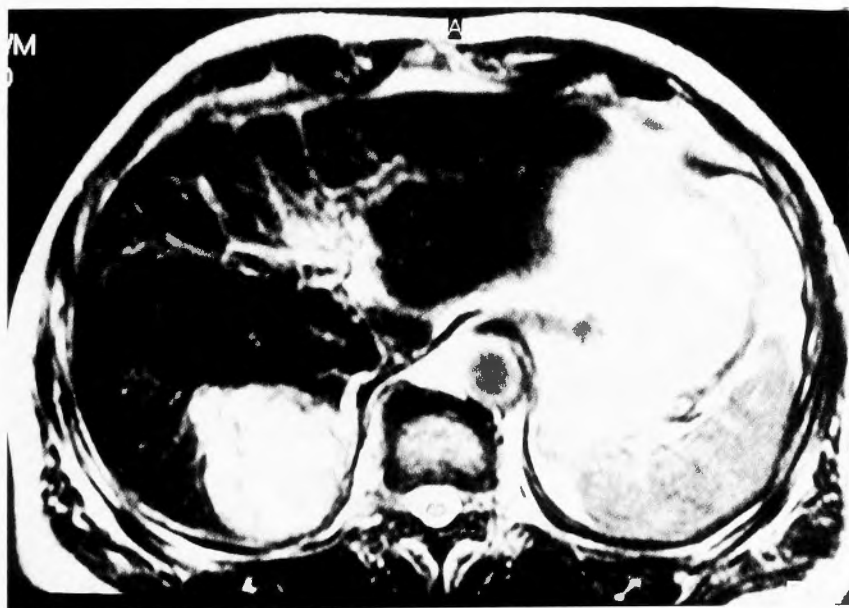


Fig. 3 Abdominal MRI findings
A high intensity region was seen on T2-weighted images. The center showed even higher signal intensity.



Fig. 4 Abdominal angiographic findings. In the venous phase, a tumor with a darkly stained margin was seen in S6.

ing in a storiform-pleomorphic type pattern. It consisted of a region showing dense proliferation of tumor cells with inflammatory cell infiltration and a region showing sparse intrastromal tumor cells, partially associated with mucinous degeneration (Fig. 6). On immunohistochemical studies of the tumor, the tumor cells stained positively for CD6 (Fig. 7) and negatively for α -fetoprotein, vimen-

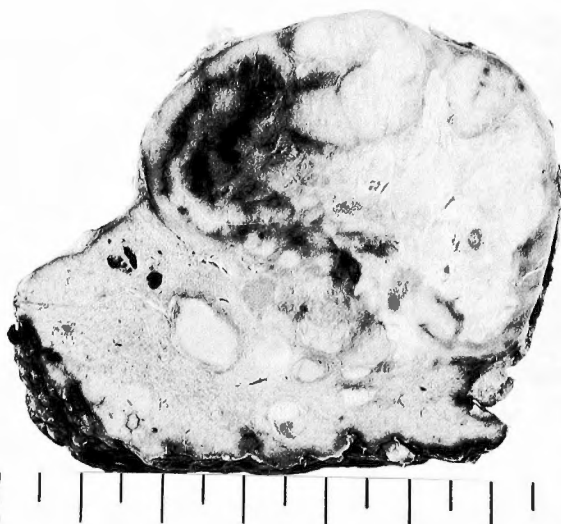


Fig. 5 Cut surface of the resected specimen. The cut surface was grayish white and parenchymatous. The border with the surrounding liver tissue showed a fibrous pseudomembrane. The tumor contained sporadic foci with hemorrhage and necrosis.

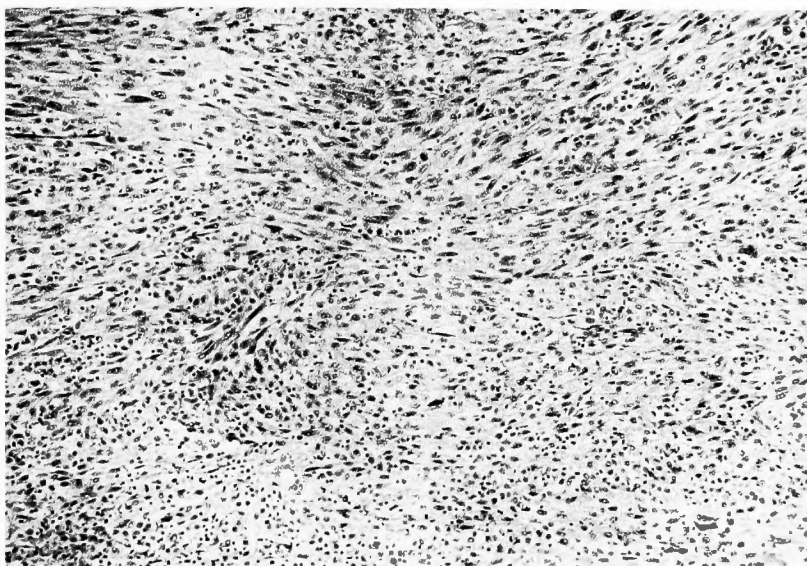


Fig. 6 Histopathological finding. The tumor consisted of elongated, spindle-shaped tumor cells proliferating in a storiform-pleomorphic type pattern.

tin, α -1-antitrypsin, α -1-antichymotrypsin, isozyme, desmin, keratin, S-100 protein, and myosin. Based on the findings described above, a primary storiform-pleomorphic type MFH of the liver was diagnosed.

The patient recovered satisfactorily after surgery and was discharged on the 31st hospital day. A chest radiograph obtained 3 months postoperatively showed evidence of multiple metastases in

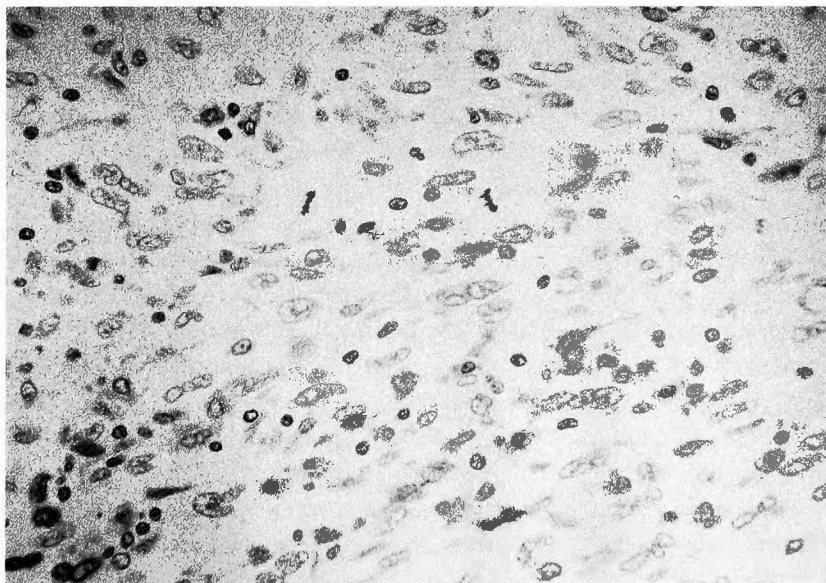


Fig. 7 Immunohistochemical findings. CD68 staining was positive.

both lung fields. He died of respiratory failure 4.5 months after the operation.

Discussion

MFH was first described by O'Brien et al. (1) in 1964. This tumor has been proposed to be of histiocytic origin (2) or undifferentiated mesenchymal cell origin (3). Since the development of immunostaining techniques, the undifferentiated mesenchymal cell theory seems more plausible. Histologically, MFH is classified into five types: (1) storiform-pleomorphic type, (2) myxoid type, (3) giant cell type, (4) inflammatory type, and (5) angiomatoid type (4). The tumor in our patient was classified as storiform-pleomorphic type, but its histologic characteristics were diverse, associated with inflammatory cells and mucous cells.

MFH generally arises in soft tissue in adults and elderly individuals and is particularly frequent on the central side of the extremities. Wiss et al. (5) reported that MFH is associated with a high incidence of local recurrence and metastasis. Local recurrence generally occurs within 2 years, and metastases are predominantly to the lung, followed by the lymph nodes, liver, and bone.

Cases of MFH arising in the liver were reported by Alberti-Flor et al. (6) in 1985 and by Nakazawa et al. in Japan in 1985 (7). To our knowledge, 16 cases of primary MFH of the liver, including ours, have been reported in Japan (7-21) (Table 2). The ages of the patients ranged from 20 to 79 years (mean, 58.2 years), and men slightly outnumbered women (10 vs. 6). Clinically, there were no characteristic symptoms. As for tumor markers, CA19-9 was elevated in 2 patients (7, 8) but no case was positive for α -fetoprotein. HBs-Ag and HCV-Ab were negative in all cases for which these variables were evaluated, whereas HCV-Ab was positive in our patient. The tumor was located in the right lobe in 11 patients and in the left lobe in 5. Tumors ranged in diameter from 4 to 18 cm and were solitary in most cases. Intrahepatic metastases were present in 3 patients, including ours (9, 10). Imaging studies characteristically revealed a hypoechoic abdominal echo. CT scans showed a low density area in the center, with ring enhancement at the tumor margin in most cases. In our patient, MRI showed a central high intensity area on T2-weighted images, suggesting the presence of necrotic foci in the center of the tumor. Angiography revealed diverse findings, including avascular and hypervascular regions, and many cases seemed to have a hypervascular region at the tumor margin. MFH was not diagnosed on the basis of preoperative imaging studies in any patient, and operation was performed for suspicion of hepatocellular carcinoma or cholangioma. The diagnosis was definitely established on histopathological examination of resected specimens. Histopathologically, all reported cases, including ours, showed a storiform pattern. Although many reported cases stained positively for vimentin and 1-antitrypsin on immunohistochemical studies, the expression of these antigens is nonspecific and is seen in various types of tumors. Our case did not stain positively for these antigens, but stained positively for CD68 (PG-M1 or KP-1). Binder et al. (22) reported that CD68, a monoclonal antibody for epitope glycoprotein antibody contained in histiocytes or macrophages, is useful in the diagnosis of MFH. Therefore, even if vimentin and 1-antitrypsin are negative, positive staining for CD68 permits the immunohistochemical diagnosis of MFH derived from mesenchymal cells of the liver including Kupffer cells.

Of the 16 cases of MFH reported in Japan, resection was performed in 14 (87.5%). However, the outcome has been very poor. Fukuyama et al. (8) reported a mean duration of survival of 11.3 months, with maximum survival of 4 years. Since effective chemotherapy is not currently available, extensive liver resection with lymph node dissection is the treatment of choice.

Table 2 Reported case of malignant fibrous histiocytoma of the liver

Author(s) (Reference)	Age Sex	Lobe Tumor(size cm)	Echogram	C T	Angiogram	Histology	Treatment	Prognose
Nakazawa et al ⁷⁾ (1985)	61/F	Right 12 x 14 x 18	hypo-hyper echoic	low density area	fine tumor vessels	storiform	r-lobectomy	
Fukuyama et al ⁸⁾ (1986)	38/F	Left 6 x 6 x 7	—	low density area	fine tumor vessels	storiform	l-lobectomy	4 years alive
Honda et al ⁹⁾ (1988)	71/F	Right	hypoechoic cystic component	low density area	fine tumor vessels	storiform/ pleomorphic	TAE	5 months died
Fujita et al ¹⁰⁾ (1988)	70/F	Left 13 x 13	—	—	—	storiform/ pleomorphic	—	3.5 months died
Katsuda et al ¹¹⁾ (1988)	61/F	Right 8.5 x 8 x 8		hypodensity area	avascular	storiform/ pleomorphic	r-lobectomy	6 months died
Ochiai et al ¹²⁾ (1988)	56/M	Right 10 x 7.5	hallo sign	low density area	avascular~ hypervascular	storiform	r-lobectomy	2 months died
Oyama et al ¹³⁾ (1989)	64/M	Right 7.4 x 7.4		low density area	avascular	storiform/ pleomorphic	r-lobectomy	142 days died
Nakajima et al ¹⁴⁾ (1990)	54/M	Right 10 x 10 x 7.5	hyper and hypoechoic	low density area	avascular	storiform/ pleomorphic	r-lobectomy	60 days died
Kono et al ¹⁵⁾ (1991)	59/M	Right 4 x 3	hyper and hypoechoic	low density area	avascular	storiform	r-lobectomy	—
Hamasaki et al ¹⁶⁾ (1991)	35/M	Left 7.5 x 6.7 x 9.1	hypoechoic	low density area	fine tumor vessels	storiform	l-lobectomy	34 months died
Akifuji et al ¹⁷⁾ (1992)	79/M	Left 8 x 8 x 6	hypoechoic	hypodensity area	avascular	storiform/ pleomorphic	l-lobectomy	Alive 8 months after operation metastasis(+)
Mima et al ¹⁸⁾ (1992)	66/M	Right 15 x 12 x 8.5	hyper and hypoechoic	hypodensity area	avascular~ hypervascular	storiform	r-lobectomy	5 days died
Nakamura et al ¹⁹⁾ (1994)	59/M	Right 4 x 3	hypoechoic	low density area	avascular	storiform	r-lobectomy	—
Kunisaki et al ²⁰⁾ (1994)	73/M	Left some nodules of 2x3x1.5 cm size	hypoechoic	low density area	—	storiform/ pleomorphic	l-lobectomy	168 days died
Iwata et al ²¹⁾ (1995)	20/M	Right 5 x 4.9	hypoechoic	low density area	avascular	storiform	l-partial lobectomy	18 months died
Present case	68/M	Right 5.5 x 6 x 6	hypoechoic	low density area	avascular~ hypervascular	storiform/ pleomorphic	r-partial lobectomy	4.5 months died

Concluding Statements

We performed liver resection in a patient who had a preoperative diagnosis of hepatocellular carcinoma. The histopathological diagnosis after operation was MFH. Patients with MFH arising in the liver who were treated surgically and cases of MFH reported in Japan were reviewed.

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和文抄録

肝原発性悪性線維組織球腫の 1 例

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江口 正信, 松本 道男

肝原発の悪性線維性組織球 (MFH) はまれな疾患である。患者は68歳の男性。主訴は倦怠感。入院時検査では血液検査で異常を認めなかった。腫瘍は肝右葉 S6 にあり大きさは $6.0 \times 6.0 \times 5.5$ cm 大であった。腹部 CT 検査で辺縁部は不均一な造影効果を伴う被膜状のものを認め、内部は不均一な low density area であった。MRI 検査では T1 強調画像で low intensity, T2 強調画像で high intensity として描出された。腹部血

管造影は、辺縁が静脈相で濃染される腫瘍像を認めた。S6 と S7 の一部を含む肝部分切除を施行した。病理組織学的検査は、線維芽細胞様細胞と組織球様細胞とからなる特徴的な storiform pattern (花むしろ模様) が見られ、腫瘍内には炎症性細胞浸潤や一部粘液変性を伴う storiform pleomorphic type (花むしろ多形型) であった。免疫組織化学的検査は CD68 に陽性を示したことより MFH と診断した。